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# Outer diameter measured by 3D CISS MRI and quasi-Moyamoya disease

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The interesting article by Kaku Y et al.: Outer diameter narrowing of the internal carotid artery and the middle cerebral arteries detected in Moyamoya disease detected on 3D constructive interference in steady-state (CISS) MR image..., shows us once more that:

1. The pathology of Moyamoya disease, especially its pathological vessel walls, compared with those of other cerebrovascular diseases like atherosclerosis.

One has to observe not only changes of the inner diameter of the pathological vessels but also that of the outer diameter caused by remodeling of vascular wall originating in the pathological process of a disease. Until now, we have paid attention only to the inner diameter, namely luminal stenosis or occlusion on the basis of angiographical or MRI findings. The 3D-CISS MRI has been revealed to be a good tool for measurement of the outer diameter to observe the latter process for instance changes of vascular wall thickness due to its remodeling.

2. The definition of Moyamoya disease and its differentiation from related diseases.

The terminology Quasi-Moyamoya disease (<ru> Moyamoya disease in Japanese) could be found, so far as the author has searched around, initially in a chapter of the book Moyamoya disease edited by Prof. J Suzuki (one of finders of this disease and has coined the name of “Moyamoya”) in 1983 [3]. Watanabe T and Suzuki N classified as “ru” Moyamoya disease which was however out of diagnostic criteria of Moyamoya disease, including:

a. Only one side corresponds with the criteria of Moyamoya disease (1) but the other side having normal vasculatures.

b. Cases with one side or bilateral middle cerebral artery MCA stenosis at the origin associated with Moyamoya vessels, however, combined with neither internal carotid artery ICA nor anterior cerebral artery ACA stenoses

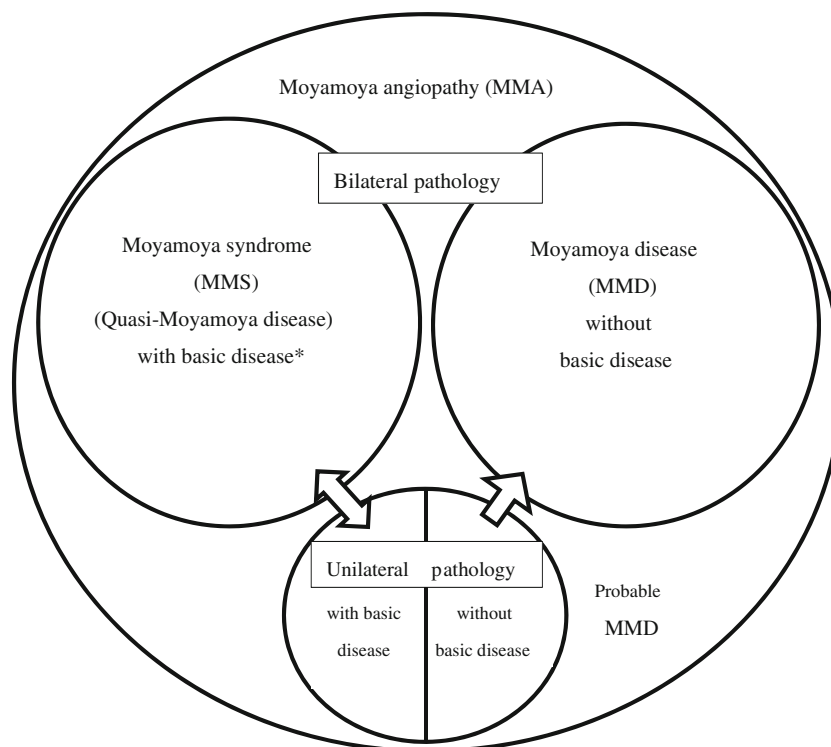
c. Combination with congenital malformation: AVM, primitive trigeminal artery

d. Associated with apparent basic disease such as: von Recklinghausen disease, head trauma (mechanical vasoconstriction), fibromuscular dysplasia, tuberculous meningitis, atherosclerosis, post-irradiation, Fanconi’s anemia, sickle cell anemia, nonspecific arteritis, connective tissue anomaly, cerebral angiitis due to leptospirosis

As you can follow, the definition of “ru” Moyamoya disease thus inherits some confusion, while the definition of the research committee under the guideline (bilaterality of findings of seno-occlusion at the terminal portion of the ICA and at proximal portion of the MCA and ACA combined with abnormal vasculatures Moyamoya at basal portion of the brain, and without any basic disease [1]) originally set in 1979 was far more clear in separating Moyamoya disease from Moyamoya syndrome having basic disease [1]. The name “ru” seems to have changed into “quasi” at the time of the English edition of the above-mentioned book in 1986 published by Springer.

The author of this editorial did not know, however, until quite recently that the research committee has now included the terminology quasi-Moyamoya disease into the diagnostic guideline in 2012 (originally in 2009 in Japanese) [2], so that this terminology can be used equally to Moyamoya syndrome, which means that quasi-Moyamoya disease also includes atherosclerosis but excludes unilateral pathology without basic disease (above mentioned in “a”) namely probable Moyamoya disease, and quasi-Moyamoya disease also includes the

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**Fig. 1** Moyamoya disease, Moyamoya syndrome (quasi-Moyamoya disease) and Moyamoya angiopathy \*basic disease [1, 2]: atherosclerosis, autoimmune disease (systemic lupus erythematosus, antiphospholipid antibody syndrome, periarteritis nodosa and Sjögren's syndrome), meningitis, von Recklinghausen's disease, brain tumors, Down's syndrome, head injury, irradiation, hyperthyroidism, stenoccephaly, Turner's syndrome, Alagille syndrome, William's syndrome, Noonan's syndrome, Marfan's syndrome, tuberous sclerosis,

Hirschsprung's disease, glycogen storage disease type 1, Prader-Willi syndrome, Wilms tumor, primary oxalosis, sickle cell disease, Fanconi's anemia, spherocytosis, eosinophilic granuloma, type II plasminogen deficiency, leptospirosis, pyruvate kinase deficiency, protein C deficiency, fibromuscular hyperplasia, osteogenesis imperfecta, polycystic kidney, oral contraceptives, and drug poisoning (cocaine, etc.)

one with hyperthyroidism as shown in Fig. 1 and attached legends. The guideline excludes, by the way, AVMs, aneurysms, or congenital vascular anomalies such as primitive trigeminal artery as “basic” disease, what was still not the case at that time in the above-mentioned original paper of “rui” Moyamoya disease, as can be seen in “c”. Differentiation of “basic disease or underlying disease” from “concomitant or associated disease” is not always easy. After all, terminology of quasi-Moyamoya disease is considered still not to be free of confusion and not to have obtained general acceptance.

The original manuscript by Kaku et al. did not include any mention of quasi-Moyamoya disease, so that it was rather clear cut in differentiating findings of 3D CISS between Moyamoya disease and atherosclerotic cerebrovascular disease. Readers are, therefore, asked not to be confused with the final version, mentioning additionally about hyperthyroidism, whose combination belongs also to quasi-Moyamoya disease or Moyamoya syndrome, which includes also atherosclerosis as basic disease.

By the way, we also use from time to time the terminology Moyamoya angiopathy MMA [4, 5], in which all the clinico-pathology of Moyamoya disease, Moyamoya syndrome, and unilateral pathology are included. This concept has originated from the idea that the management–treatment of ischemia, epilepsy, or bleeding due to this entity can be performed on the base of similar clinico-pathophysiology and not necessarily based on the etiology.

**Conflict of interest** None.

## References

1. Fukui M (1997) Members on the research committee on spontaneous occlusion of the circle of Willis (Moyamoya disease) of the ministry of Health and Welfare, Japan: Guidelines for the diagnosis and treatment of spontaneous occlusion of the circle of Willis (“moyamoya” disease). Clin Neurol Neurosurg 99(suppl 2):S238–S240

2. Research committee on the pathology and treatment of spontaneous occlusion of the circle of Willis (2012) Guidelines for diagnosis and treatment of Moyamoya disease (Spontaneous occlusion of the circle of Willis). *Neurol Med Chir (Tokyo)* 52:245–266
3. Watanabe T, Suzuki N: “Rui”-Moyamoya (1983) in Suzuki J(ed): Moyamoya disease. Tokyo, Igaku-Shoin, pp 123–145 (Japan)
4. Yonekawa Y, Takagi Y, Khan N<sup>⊗</sup> (2011) Adult Moyamoya disease. In: Winn HR (ed) *Youmans Neurological Surgery*. Elsevier, Vol 4, Chapter 356, pp 3689–3697
5. Yonekawa Y, Operative neurosurgery (2012) Personal view and historical backgrounds (9) Moyamoya angiopathy (MMA): Past history and status presens. *No Sjinkei Geka* 40(1):67–87, in Jpn, Eng abstract

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